

PULMONARY ARTERY ATRESIA**Healthcare**

Keywords: pulmonary atresia, balloon atrioseptostomy, Blalock-Taussigshuntss.

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Abstract

Introuction: Pulmonary atresia is a congenital malformation of the heart, valve in which the pulmonary valve orifice fails to develop. The valve is completely closed thereby obstructing the outflow of blood from the heart to the lungs. Doctors are unsure of the cause of congenital heart defects, but there are some medical conditions that have been found to increase the risk of having a baby with a heart defect such as congenital heart disease in the mother, father, brother, or sister of the baby, a diabetic mother, use of drugs or alcohol or over the counter prescriptions during pregnancy. There are many different types of classification of PA but more useful is classification in PA with and without interventricular defect. Aim of presentation: is to present neonates with pulmonary atresia, with or without interventricular defect in group of children with central cyanosis and possibility to make diagnosis of PA with echocardiographic examinations. **Methodology:** Retrospectively we analyzed children with central cyanosis where etiology of cyanosis where any congenital heart defect. **Results:** During the period 2008 – 2013 in Pediatric Clinic in Prishtina 28 neonates with PA were diagnosed. Of them 19 were with interventricular defect and 9 without. In group of neonates with PA and without DIV communication at the atrial level was restrictive in 8 of them and in 4 balloon atrioseptostomy procedure was successfully performed. 5 of remaining has died, in absent of the possibility for palliative surgical intervention.

Introduction

Obstructive changes of output part of the right ventricle can be presented in every part of it whereas the rate of obstruction ranges from a slight obstruction to full atresia of the pulmonary artery. The most frequent localization of obstruction is at the level of pulmonary artery, but it can be found in the other parts such as infundibulum or at the distal part of the pulmonary artery. The most severe form of obstruction is the atresia of PA, localized at the level of the valves.¹ To simplify this very complicated and large problem, there is a classification of different forms of the PA.

According to this, PA can be classified in 3 forms:

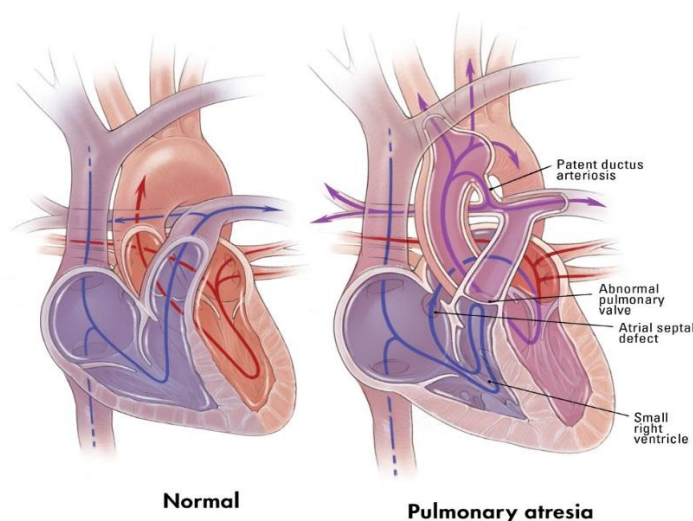
- PA atresia with ventricular septal defect (PA/VSD)
- PA atresia with intact ventricular septum (PA/IVS)
- PA atresia with complex malformations such as interventricular defect, double inlet atrioventricular connections, with corrected or complete transposition.³

Pulmonary atresia with ventricular septal defect (PA/VSD)

It can be said that this entity represents the most severe form of tetralogy of Fallot although in hemodynamic and anatomic aspect in this malformation exist differences of tetralogy by the way of blood supply to the arterial tree, which regularly is of aorta, in retrograde way. This malformation is accompanied by lots of pathoanatomical and hemodynamic changes which often are complicated and in need of a good presentation, invasive research is needed.⁷ Clinical manifestations: In many elements PA is similar to the clinical picture of the severe form of tetralogy of Fallot. Central cyanosis dominates as clinical sign which comes on immediately after birth, whereas the cyanosis degree depends on number and diameter of collaterals which supply the pulmonary circulation. Collaterals are not sufficient at the largest number of children, so the cyanosis deteriorate more in case of arterial duct closure. There is no sound at the infundibular level, but there is no sound at VSD too, because of the pressure balance between ventricles.⁸

Pulmonary atresia with intact ventricular septum (PA/IVS)

All of the morphological and hemodynamic changes of this abnormality are similar to those that are described to the first malformation, interventricular septal continuity is the only difference. This abnormality is not just a imperforated membrane at the pulmonary artery level, but regularly is presented as a complex malformation in which dominates the obstruction of the outlet place of the right ventricle. The right ventricle and tricuspidal valve are abnormal in some cases like: hypoplastic right ventricle, infundibulum atresia, tricuspidal valve dysplasia or hypoplasia etc. Rarely the changes of tricuspidal valve looks like the changes of Ebstein malformation.^{9,10} The treatment of kids with PA still is a big problem and a big challenge in cardiopediatry. In modern medicine it means the birth planning, treatment of a newborn with the right therapy (Prostaglandine) and preparation for palliative or definitive early intervention, depending on the presentation form and pathoanatomy.¹¹



Cardiogram 1. Pulmonary artery atresia

Incidence: Incidence: This abnormality participates with 1-3% in all congenital heart malformations whereas 1/4 of child and neonatal age with central cyanosis belong to this entity. Freedoom and Keith, in a large series of children diagnosed with *AKZ* in Toronto, report an incidence of 2.5%, with slight predominance of males. In another study at Children's Hospital in Pittsburgh reported a higher incidence of 2.4% with the AP atresia of all the children who have the need for surgical intervention.

Results

In the group of 314 children analyzed in our study under the age of 28 days, with central cyanosis, 28 children (8.9%) belong to this entity. Of these 19 children (67.85%) were male and 9 children (32.15%) are women. This high incidence in our group does not correspond to the statistical data and argued that in our study is about a selected pathology. These criteria include age of children (in our study only analyzed congenital malformations who in this age manifested with central cyanosis in the neonatal age) and group of children from all territory of Kosovo with close clinical specifications.

Malformations	D-TGA	TOF	DORV	Atresio AP	Sy HLV	complex AKZ	TAPVR	Atresio VT	EA
Child number	106	96	42	28	14	11	9	6	2
%	33.75	30.57	13.37	8.91	4.45	3.5	2.86	1.91	0.63

Table 1. The incidence of complex malformation in the studied group

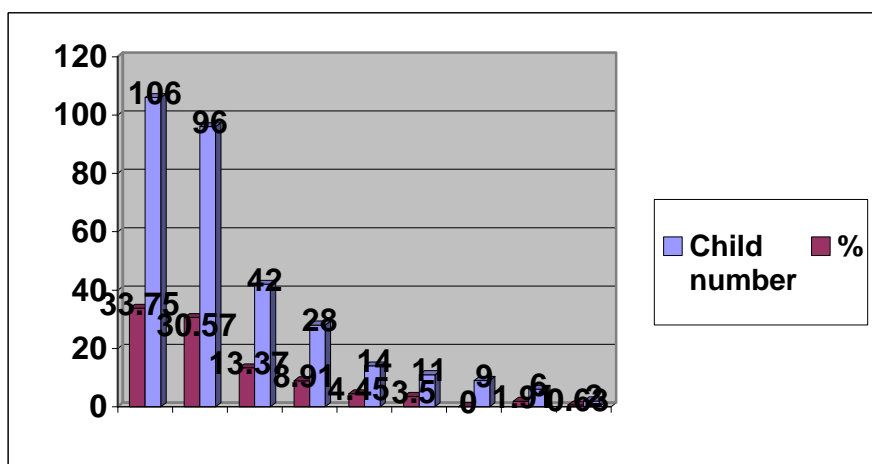


Fig.1. The incidence of complex mal-formations in our group of study

Child number with CQ	1283
Child numb. With AKZ	314
Child numb. With PA	28 13.37 %

Table 2. The number of children with PA atresia compared to the children with CQ and KLZ

Discussion

Pathoanatomic changes to PA atresia without VSD

By definition it shows that the outlet part and the pulmonary valve are atretic, whereas interventricular septum is intact. The distal arterial trunk and branches of the AP can be well developed and more rarely are dysplastic or may be, the most severe form, atretic. Changes in this section do not correspond to changes in the right ventricle. Right ventricle of heart happen to be small, to be associated with arterial trunk normal size. In pathoanatomic aspect right ventricle changes can be divided into two entities:

- Commissural crist is at the central part pulmonary artery root. This form is accompanied with infundibular atresia or severe tricuspid regurgitation as main Ebstein malformation.
- The second option is with peripheral circles but central part is polished and shaped dome. This form is associated with the presence of infundibulum and high pressure in the right ventricle, which develops at the right ventricular outflow.^{12,13}

In our group 20 children (71.42%) had a first form changes and 8 children (28.57%) were with second form changes.

Right ventricular space is hypoplastic but data from literature has shown that right ventricular spaces can have a wide range of morphologies. In this respect is proposed the measurement of several parameters within the right ventricle, particularly Inlet and outlet portion dimensions, and these values to be compared with the values of the left ventricle. The largest number of children have lower values of the right ventricle, a number of children have normal values and rarely have greater value.^{13,14,15} In our study we did not assess on the basis of measurements but based on visual values and are of the opinion that most of the children had right ventricular hypoplasia, rarely it has been normal values and right ventricular walls regularly were hypertrophic.

Tricuspid valve apparatus corresponds closely with right ventricular changes. In the majority of cases tricuspid valve and tricuspid annulus are small. Changes can be seen also in dimensions, form and function of tricuspid valve blades.

Even in our study, aortic and tricuspid valve values visually correspond to the values of the right ventricle. Regularly tricuspid valve function has been compromised and is manifested by regurgitation of easy scale up to more severe forms, by imitating the Ebstein's anomaly.^{16,17}

Coronary artery malformations are common with this anomaly and are high risk factor for definitive prognosis. Fistula communications between the right ventricle and coronary arteries, according to publications are seen to 1/3 of children with PA atresia, especially to right ventricle hypoplasia form. It is believed that the basis of the large number of coronary-ventricular fistulas is high pressure which reigns in right ventricular space and persistence of right ventricular communications and coronary circulation during embryology. Changes in coronary arteries can also be in terms of lumen diameter of coronary arteries, showing narrow forms, obstruction or dilatation part with them.¹⁸

In our group of 28 children, with Doppler, concluded expansion of proximal part of the right coronary artery at 4 children (14:28%). In right ventricular space of these children, Doppler recorded turbulent flow during the systole which is characteristic for coronary-caval fistula. Later, to the three children from this group, have been recorded presence of coronary fistulas during cardiac intervention.

Pulmonary-systemic and duct collaterals are rare. Coronary arteries malformations are frequent, interatrial septum defect is required for survival of these children.

Clinical-Laboratorial Examinations of PA Atresia

Electrocardiography (ECG)

ECG is regularly abnormal and reflects the morphological changes in the heart. The pace and the axis are normal, there are signs of left ventricular hypertrophy or biventricular hypertrophy. P wave dominates as a result of load on the right ventricle. Signs of myocardial ischemia manifest in the presence of coronary anomalies.¹⁹

All children in our group have signs of changes in the ECG. All children registered sinus rhythm and P wave is dominant only in 9 children, who exhibited severe tricuspid regurgitation. All children in our group have different degree of deviation to the right axis. No children exhibited any rhythm disorders. The 9 children who exhibited aortopulmonary fistula, have been recorded the changes in ST segment depression.

Heart teloradiography

All children in the heart teloradiography have similar results to those of literature consulted in terms of teloradiography. Slight oligemia is regularly present.

Echocardiographic examination

In all centers in the world and in our study Echocardiographic examinations and findings are the fundamental basis of this anomaly diagnosis, especially in the examination of the right ventricular morphology. Echocardiographic diagnosis can be made to the ante and postnatal age.

Antenatal echocardiography

From the 14th week of gestational age can be diagnosed the right ventricular outflow atresia and the accompanying anomalies. With the use of modern apparatus with Doppler and color-Doppler, clearly presented pathoanatomia of misinformation from the part of right ventricular outflow, size and right ventricular dimensions and anatomy of the tricuspid valve apparatus. Early registration of normal dimensions right ventricle and regurgitation at tricuspid valve level are signs of high risk and poor prognosis of abnormality.^{19, 20}

Despite anamnestic data from mothers, for regular echosonography examinations during pregnancy of any children in our group has not been diagnosed in the neonatal age.

Postnatal echosonography

In classical cutting, 2-D echocardiography provides comprehensive presentation of morphology and hemodynamically disorders, presentation of coronary artery anomalies and possible collateral. Today, the children with restrictive interatrial communication, with echocardiographic tracking is made the balloon atrioseptostomy intervention - Rashkind's procedures. In detail is analyzed the morphology of the tricuspid valve, measuring its dimensions, regurgitation and eventual dysplasia of tricuspid valve blades. Analyses anterograde circulation and regurgitation rate, under which is assessed the pressure in the right ventricle. Right ventricle of heart and part of his output are part of special examinations and ways of supplying blood to the pulmonary artery. Assessed hemodynamic in arterial duct and eventual aortopulmonary collaterals.^{18, 19, 21}

Hemodynamic changes which take place and are specific to this anomaly clearly are presented with Doppler and color-Doppler and, at the same time this method of examination today is sufficient and completely eliminates the need for invasive examinations.

In all children on our group diagnosis of pulmonary artery atresia was decided after echocardiographic examination. In interatrial communication assessment for 22 children (78.57%) in neonatal age it is regarded as sufficient. In 6 children (21.42%) communication has been restrictive whereas their bad health condition at the time of examination, has not allowed the balloon atrioseptostomy intervention. All the children at restrictive interatrial communication in our group have died in the early neonatal age.

During the echocardiographic examination we have paid particular significance to tricuspid valve apparatus by presenting the anulus, tricuspid valve blades, anterograde flow and tricuspid regurgitation. The regurgitation degree at the tricuspid level, in our group also corresponds with that of the literature references. All the children with severe tricuspid regurgitation in our group have early developed congestive insufficiency of the right ventricle and in long monitoring, only 18 children (64.28%) have reached the age of 6 months. In all of these children aortopulmonary collaterals are also important.

With echocardiographic examination are presented the morphology of the right ventricle, wall hypertrophy and also the plase and degree of obstruction. Particular attention has been paid to the pulmonary artery trunk and branches as well as the way of the blood supply to the pulmonary circulation. In 12 children (42.85%) arterial duct has an uncharacteristic anatomy: in 6 children it has descended from subcalvia artery dexter and is tortuouse form. In 21 children (75%) are registered aortopulmonary communications.

Meanwhile, after examining echocardiographic data obtained, cardiology and cardiosurgery consultations, is planned one of palliative surgical procedures (according Kirklin's or its modifications), which today are made in different age child, preparing the child for definitive subsequent intervention.

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